LETTERS TO THE EDITOR

Regarding "Surgical treatment of venous malformations in Klippel-Trénaunay syndrome"

To the Editors:

We read with interest the excellent article by Noel et al (J Vasc Surg 2000;32:840-6), dealing with the preoperative evaluation of Klippel-Trénaunay syndrome (KTS).

The syndrome is relatively rare and complex, requiring a multidisciplinary approach in a specialized vascular center of excellence to obtain successful management.[1,2]

We would like to discuss certain points of this article.

1. The classic triad of capillary malformations, limb hypertrophy, and atypical local varicosities is not always present. In cases when only two of the above findings are present, associated with other dysplasias such as lymph vessel anomalies, mitral valve prolapse, or spinal stenosis (consisting of a multisystem disease), could we consider an atypical presentation of KTS?
2. Is polydactyly considered hypertrophy or hyperplasia? Or is it a manifestation of the syndrome for which, like its pathogenesis, very little is known? We certainly agree with the authors that soft tissue and bone hypertrophy are not the direct "consequences of venous stasis," because a supernumerary toe cannot be explained without the presence of acromegaly.
3. Do venous ulcers represent an evolution in the natural history of the syndrome, or are they due to chronic venous insufficiency, as in cases without congenital anomalies of the deep venous system? And how can the need for stripping of the great saphenous vein be explained apart from dealing only with the laterally localized varicosities?
4. There are reports in the literature[3] that in some cases of KTS there are no macrofistula arteriovenous communications. Could the presence of high intraluminal pressures, the macroscopically rose-colored venous blood and the increased oxygenation, be explained by the presence of microfistula arteriovenous communications?
5. The authors reported good function with vein patency 102 months following popliteal to saphenous deep vein reconstruction. Since no transient arteriovenous communication was used, the good result can be attributed only to good technique and anticoagulation or to an increased intraluminal pressure gradient. Could the latter be responsible for the high incidence of recurrence (50%)? A comparative study of the intraluminal pressures preoperatively and postoperatively would be interesting.

We would like to congratulate the authors for their thorough preoperative, morphological, and functional study of their patients. We would also like to report the case of a 60-year-old patient who during childhood had had an above-knee amputation because of edema and gangrene of the lower leg after varicose vein surgery.[4] The patient exhibited scars on the lateral surface of the stump, hemangiomas, and recurrent varicosities. Phlebography in our institution showed complete agenesis of the deep venous system, which explains the bad outcome of his operative treatment.

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REFERENCES

1. Dimakakos PB, Katsenis K, Arapoglou V, Papasava M, Antoniou A. Therapeutic management and results of congenital angiodysplasias. Phlebology 1995;1(Suppl):1003-5.
2. Dimakakos PB, Papageorgiou A, Mourikis D, Kotsis TH, Papadimitriou J. Therapeutic tactics and late results in predominant truncal congenital malformation. J Cardiovasc Surg 2000;41:447-55.
3. Szilagyi DE, Smith RF, Elliott JP, Hageman JH. Congenital arteriovenous anomalies of the limbs. Arch Surg 1976;111:423-9.
4. Dimakakos PB, Androulakakis Z, Arapoglou V, Papadimitriou J. Management of congenital vascular malformations of the limbs. Int J Angiol 1994;3:26-31.

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Reply

Drs Dimakakos and Portinos raise several issues regarding the complex clinical presentation and treatment of Klippel-Trénaunay syndrome (KTS). The presence of the classic triad of limb hypertrophy, varicosity, and capillary malformation (port-wine stain) is not always present. In the Mayo Clinic series, published by Jacob et al,[1] only 159 (63%) of 246 patients had all three features of the syndrome. Lymphatic anomalies have also been noted in KTS, and they are probably much more frequent than reported. Limb hypertrophy and swelling and the mixed vascular malformations in the subcutaneous tissues and muscles almost always have a component of lymphatic abnormality. The frequency of digital anomalies is well-known, and an excellent article on this has been published in the orthopedic literature by Amadio et al.[2] Syndactyly and polydactyly have been the most frequent digital anomalies in our recent review.[1] The pathogenesis of such skeletal anomalies is unknown, but it is very likely that the soft tissue and bone hypertrophy are not direct consequences of the hemodynamic changes of the vascular malformations but are independent defects in the limb development. Concomitant mitral valve prolapse or spinal stenosis, mentioned by the authors, are rare.

Venous ulcers are usually results of venous hypertension, although erosion of the skin can also be due to extensive superficial vascular malformations. Venous hypertension should be relieved with stripping of the superficial incompetent veins, removal of incompetent avalvular superficial embryonic veins, varicose vein avulsion, or subfascial endoscopic perforator vein ligation, when necessary. The authors are right that the saphenous vein is rarely involved and if competent, should not be stripped. Occasionally, however, the greater saphenous vein is also enlarged and incompetent, and if the deep system is patent, the saphenous vein in these patients can be removed, with good clinical results. We cannot emphasize enough, however, the need for thorough preoperative imaging with contrast or magnetic resonance venography and duplex scanning.

High-flow, high-shunt arteriovenous fistulas are not present in KTS, but clinically insignificant microfistulas may occur. One theory of the etiology of KTS suggests that intrauterine injury to the sympathetic ganglia may result in microscopic arteriovenous anastomoses. The clinical significance of this, however, is unknown.

When considering deep venous reconstruction in KTS, preoperative pressure measurements, as suggested by the authors, are helpful to confirm that collateral circulation is inadequate. The

larger the pressure gradient, the better is the chance of graft patency. Venous bypasses in KT patients are rarely needed. Most patients, as mentioned in the manuscript, are managed conservatively. We must reiterate, as the authors did in their letter, the need for multidisciplinary management of patients with KTS in specialized vascular center.

We thank Drs Dimakakos and Portinos for considering these salient points and for their active interest in this fascinating disease.

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REFERENCES

1. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel Trenaunay syndrome: spectrum and management. Mayo Clin Proc 1998;73:28-36.
2. Amadio PC, Dobyns JH, Stickler GB, Unni KK. Anomalies of the fingers and toes associated with Klippel-Trenaunay syndrome. J Bone Joint Surg 1991;73A:1537-46.

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